Management of the Fetus with a Correctable Defect

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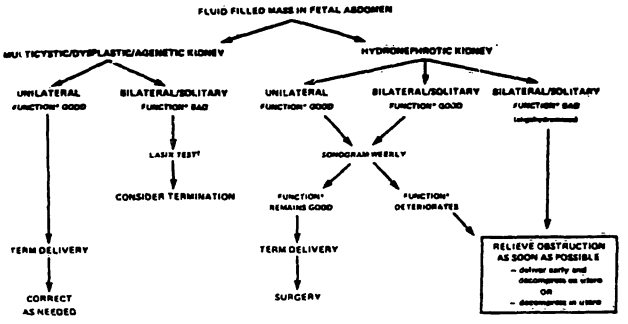
Many fetal abnormalities can now be detected. Most defects are best treated after birth, and prenatal diagnosis improves outcome by allowing the patient, family, and physician to discuss the alternatives; to choose the optimal time, mode, and place of delivery; and to prepare for optimal postnatal care.

Only a few disorders are potentially amenable to treatment before birth. Fetal deficiencies may be alleviated by the provision of a missing substance, as in intrauterine transfusion for fetal anemia. Although numerous anatomic malformations are detectable before birth, the only ones that warrant consideration for treatment in utero are simple structural defects that interfere with organ development and whose alleviation might allow fetal development to proceed normally. At present, congenital diaphragmatic hernia, hydronephrosis, and hydrocephalus are candidates. Studies in animal models have helped to define the pathophysiology of these lesions.

The most difficult problem in prenatal management is how to select only the fetuses that might benefit from treatment. Since there is a broad spectrum of severity of each lesion, careful serial assessment of fetal anatomy and organ function is necessary to exclude those so severely affected that they cannot be saved and those so mildly affected that they will do well with standard postnatal treatment. Unilateral hydronephrosis and mild bilateral hydronephrosis with evidence of good renal function do not require in utero treatment. The fetus older than 32 weeks may benefit from early delivery for immediate postnatal decompression. However, the fetus with bilateral hydronephrosis secondary to urethral obstruction in which severe oligohydramnios develops before the last trimester will predictably have pulmonary hypoplasia and renal damage incompatible with life. There is a growing experience with decompression of the obstructed urinary tract into the amniotic fluid through a catheter shunt placed percutaneously. Catheter malfunction and malposition required repeat catheterization in some cases. The outcome for fetuses with early severe oligohydramnios is discouraging, suggesting that many of these severely affected fetuses may not be salvageable even with decompression. Many neonates have survived with good renal function, but it is not certain that treatment influenced outcome in the milder cases. Prenatal decompression should be performed only when the fetus with bilateral hydronephrosis has evidence of compromised renal function (oligohydramnios or documented diminution of amniotic fluid on serial sonograms) and is too immature to be delivered for postnatal decompression. For severe cases with oligohydramnios before 22 weeks, the ability of the fetal kidney to produce urine (after bladder aspiration or drainage) should be established before more definitive treatment is considered.

Although the natural history of fetal ventriculomegaly is not well documented, a review of the available evidence suggests that a fetus with high-pressure obstructive hydrocephalus may benefit from decompression before birth. Decompression of the ventricles by percutaneously placed ventriculoamniotic-shunt catheters has decreased ventricular size, but the efficacy of treatment cannot be determined until long-term postnatal neurologic follow-up is available. Prenatal decompression should be performed only if the fetus with isolated ventriculomegaly has evidence of progressive dilatation and decreasing mantle thickness on serial sonograms, is too immature to be delivered for postnatal shunting, and has no other serious central-nervous-system abnormalities on careful sonographic screening.

Treatment of the fetus with a potentially correctable defect is promising but still experimental.



ABDOMINAL WALL DEFECTS - PRENATAL ASSESSMENT